RES MEDICA
Journal of the Royal Medical Society

Portal Hypertension

J. G. Clark

Abstract

Definition
Portal hypertension is a condition of chronically raised pressure in the portal venous system, giving rise to splenomegaly and the development of enlarged collateral venous channels connecting the portal and systemic circulatory systems.

Anatomy
Figure I illustrates the disposition of the main veins of the portal system and indicates the chief sites at which this system communicates with the systemic veins. The presence of a Caput Medusae in cases of intrahepatic obstruction, and its absence in extrahepatic obstruction. is evident from the drawing. Not all of these sites are important clinically or diagnostically, but their total capacity causes a very considerable volume of blood to bypass the liver and be released into the general circulation when the communications are enlarged.

ISSN: 2051-7580 (Online) ISSN: 0482-3206 (Print)
Res Medica is published by the Royal Medical Society, 5/5 Bristo Square, Edinburgh, EH8 9AL

Res Medica, Sumer 1960, 2(2): 13-20, 46
doi:10.2218/resmedica.v2i2.337

PORTAL HYPERTENSION

By J. G. CLARK

Based on a Dissertation read before the Royal Medical Society on Friday, 15th January 1960.

Definition

Portal hypertension is a condition of chronically raised pressure in the portal venous system, giving rise to splenomegaly and the development of enlarged collateral venous channels connecting the portal and systemic circulatory systems.

Anatomy

Figure 1 illustrates the disposition of the main veins of the portal system, and indicates the chief sites at which this system communicates with the systemic veins. The presence of a Caput Medusae in cases of intrahepatic

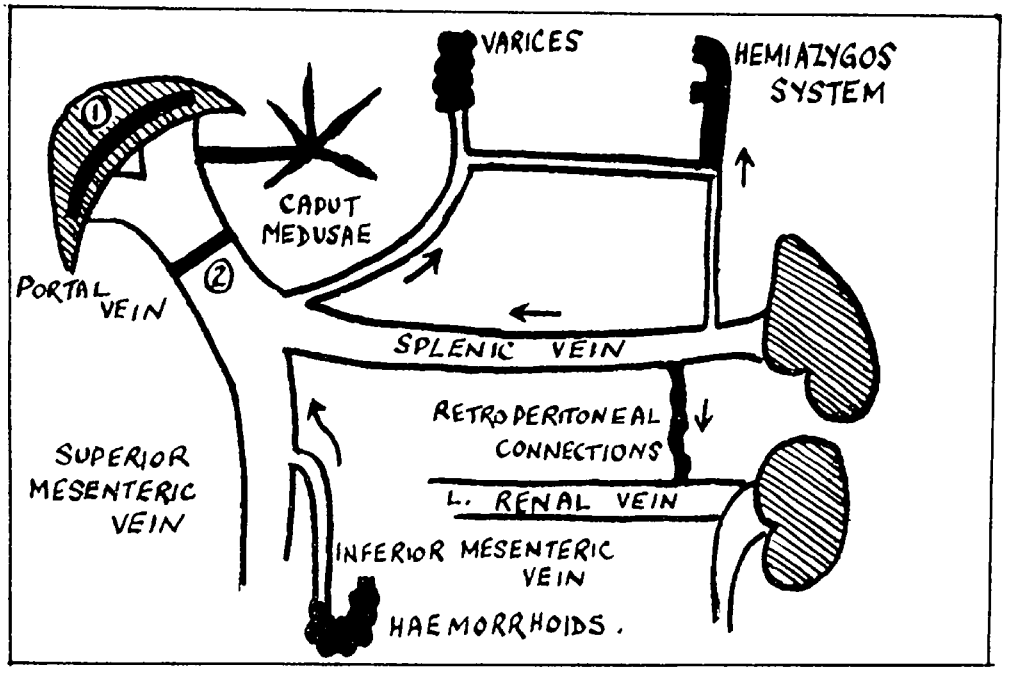

DIAGRAM OF PORTO-SYSTEMIC CONNECTIONS

① INTRAHEPATIC BLOCK

② EXTRAHEPATIC BLOCK.

Fig. 1

obstruction, and its absence in extrahepatic obstruction, is evident from the drawing. Not all of these sites are important clinically or diagnostically, but their total capacity causes a very considerable volume of blood to bypass

the liver and be released into the general circulation when the communications are enlarged.

The vascular structure of the liver is important, and I have illustrated this with a diagram (Figure 2) on which it is readily seen how easily inflammatory and fibrotic processes can cause obstruction of flow and therefore raised portal pressure. Understanding of this structure is fundamental to the comprehension of liver pathology, and the clinical conditions to which it can give rise.

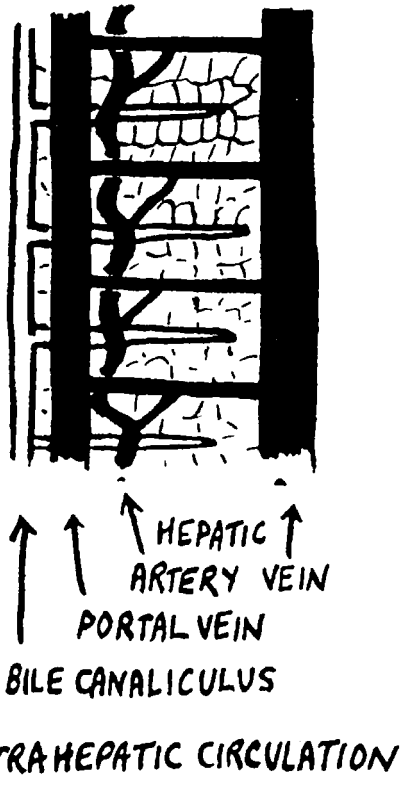

Fig. 2

Physiology

There is no justification for attempting to include here the ocean of hepatic physiology in the sea of portal hypertension, and I shall limit my remarks to three subjects.

Firstly, in the realm of biochemistry, *protein metabolism* is of greatest interest in this condition, but it will be more convenient to discuss that later on in connection with portal neuropathy.

Second, *blood flow*. The amount of blood reaching the liver is controlled by the sympathetic system operating in the gut. This variable input is dealt with in the hepatic vascular bed by a mechanism which is as yet ill-understood. It has been shown in rats that the area of liver bed in use at any one time is controlled by means of opening and closing of the peripheral parts of it. The mechanism by which this is perpetrated is not known, and the existence of porto-hepatic communications has been proposed by some workers and denied by others.

Total liver blood flow in a normal adult has been shown to be about 1600 ml./min.

Third, *portal pressure*. This can be measured in three ways:—

(*a*) Canulation of an omental vein at operation.
(*b*) Percutaneous splenic puncture.
(*c*) Wedge manometry of hepatic venous radicals.

The third method can only be done at a well equipped cardiology centre.

Normal portal pressure is 10-16 cm. of saline, as measured by omental canulation, and a little higher on splenic puncture (12-20 cm. of saline). Temporary occlusion of the portal vein in a normal subject will cause the pressure to rise to 50 cm. saline, and pressures of 20-50 cm. saline are typical of portal hypertension.

Pathology

Causal conditions may be divided into three groups:—

(*a*) Prehepatic venous occlusion, from within or without.
(*b*) Intrahepatic sinusoidal obstruction, by inflammation or fibrosis—80% of cases.
(*c*) Post-hepatic venous occlusion—very rare.

Space does not permit me to give details, which can be discovered in any text-book on pathology. The pathological changes due to portal hypertension are as follows:

(1) **Varices**—By far the most important of these are oesophageal varices, but, rarely, varices in the falciform ligament give rise to the interesting Cruveilhier-Baumgarten syndrome, and retroperitoneal varices may rupture giving a massive fatal haemoperitoneum. Haemorrhoids, for some unknown reason which apparently defies logic, are but a rare manifestation of portal obstruction. If a patient comes to you with haemorrhoids, start looking for a cause in the anorectal region, and not in the liver.

(2) **Portal vein thrombosis**—Occurs in about 13% of cases, and may give rise to spreading mesenteric thrombosis.

(3) **Splenomegaly**—This is thought to be due in part to the raised pressure, and in part to an actual hypertrophy. This latter theory is based on evidence obtained from transplantation of the spleens of rabbits under the skin of their abdominal walls, where a distinct hyperplasia was observed secondary to induced portal hypertension.

Cinical Features

These can easily be worked out from the pathology. The most important is *haemorrhage from the varices*, which may be sudden and torrential, preceded by a salty taste at the back of the mouth, or so slow that the only symptom is breathlessness due to the secondary anaemia. After the bleed the patient may pass into coma, either due to shock or due to neuropathy consequent upon the absorption of blood from the gut. Characteristically the bleeds are unpredictable, there being anything from days to years between them. The cause of bleeding has been attributed to:

(*a*) Trauma of food particles,

(*b*) Peptic ulceration (Learmonth),

(*c*) Raised intra-abdominal pressure—which may increase the portal venous pressure by as much as 130 cm. of saline, for example in coughing.

Splenomegaly, in conjunction with a heavy haematemesis, and in the absence of a "peptic" history, should make one suspicious of portal hypertension. *Hypersplenism* is the condition of splenomegaly associated with anaemia, leucopenia and thrombocytopenia. This, with the hepatomegaly which sometimes occurs concomitantly, has been referred to as "Banti's syndrome," but this term should be abandoned now that the common factor of portal hypertension is recognised. Some of the *anaemia* is thought to be due to increased erythrophagocytosis of the cells which spend an unduly long time in the splenic pulp, but also blood is lost in the form of melaena, haematemesis and epistaxis. Thrombocytopenia is the barb upon the hook of oesophageal varices. No pathological process could have been more cunning than to provide a patient with a potential source of bleeding in the form of varices and then to deprive him of part of his clotting mechanism. If an experienced clinician, enwrapped in the worship of Bacchus, is unaware of this fact, what chance has the layman?

Ascites is not a feature of hypertension *per se*, but occurs if there is associated liver damage. It has been variously ascribed to a localised manifestation of a generalised fluid retention due to failure of the liver to inactivate sodium retaining corticoids, to increased vascular permeability

consequent upon the local hypertension, and to low plasma albumin, and is more evident when the obstruction is post-sinusoidal than in pre-sinusoidal block.

Signs of hepatic insufficiency may also be present. Thus there may be jaundice, with derangement of liver function tests, red palms, pigmentation of skin creases, spider naevi, xanthomata, and muscle wasting.

The collateral circulation may make itself evident in the form of a Caput Medusae in intrahepatic obstruction, or dilated veins in the scar of an abdominal operation. As mentioned above, haemorrhoids cannot be taken as a classical sign of portal hypertension.

Lastly, there may be symptoms of vague ill health—anorexia, mild indigestion, lassitude, and sometimes an ill-defined discomfort in the right hypochondrium.

Diagnosis and Investigations

The cardinal features of this disease are splenomegaly and haematemesis. There may be nothing else in the clinical examination to guide the physician, and often splenomegaly is the only evidence presented. But there remains a battery of investigations which should not fail to elucidate the true state of affairs within the abdomen.

(1) **Liver Function Tests** may indicate that the liver is at fault, and help to decide the surgeon for or against a decompression operation (*v. infra*). It is important to realise that negative tests do not rule out liver pathology or portal hypertension.

(2) **Barium Swallow** can be relied upon to visualise the varices as chains of rounded filling defects in at least 40% of cases.

(3) **Oesophagoscopy** is suggested for doubtful cases, but I would hesitate to advise this for a patient with an acute bleed.

(4) **Splenic venography** is perhaps the most decisive and useful test available, and is indispensible to the surgeon. Originally this was carried out via an omental vein at laparotomy, but recently the technique of percutaneous transplenic venography has been developed, in which a few ccs. of opaque medium are injected into the splenic pulp, whence it is swept into the portal system which is therefore visualised. This is now a safe and effective procedure.

Only a few months ago a new approach to venography was made. This time the hemiazygos system was visualised by an intra-costal injection of opaque medium. This was claimed to be useful in difficult diagnostic problems, but its value remains to be confirmed.

(5) **Liver Biopsy** may be done only when the information it yields is going to influence the management of the patient, and when the blood clotting mechanisms have been proved satisfactory.

(6) **Peritoneoscopy** has been developed for use when malignant disease of the liver is suspected.

Treatment

Before treatment is begun the diagnosis of hypertension with normal liver function, or hypertension with damaged liver should be established. This done, treatment is aimed at

(1) Stopping the bleeding if it is occurring,

(2) Management of liver insufficiency if present,

(3) Reducing the portal pressure by surgical means.

But it must be realised that no treatment, in our present state of enlightenment, will in any way alter the course of hepatic pathology, and we can provide only temporary relief. We can guide our patient safely through the rapids, but inevitably Niagara draws nearer.

Firstly, the patient with signs of portal hypertension: the non-operative methods of reducing portal hypertension consist in:—

(*a*) A low salt diet (less than 5 grams per day).

(*b*) Diuretics, either mersalyl or chlorothiazide.

Recent workers claim that quite dramatic results can come from treatment with chlorothiazide, but we have to face up to the chronicity of this disease, and I think it would be fair to say that diuretic drugs have their place in cases of acute hypertension due to active hepatitis, during the pre-operative phase of chronic cases, and in patients for whom surgery is for some reason contra-indicated.

What can he eat? With impaired liver function and in the absence of hepatic failure, he should be given a high protein diet with plenty of carbohydrate and added glucose. He must abstain completely from alcohol, and iron should be given to help him correct his anaemia.

More often portal hypertension presents itself as an acute emergency—as a haematemesis. The immediate measures to be taken are: rest in bed, transfusion and sedation. Care must be taken with morphia, which is detoxicated in the liver, and no more than 8 mg. should be given subcutaneously. Apart from this one drawback it is an admirable drug.

Having set up a blood drip, or plasma if there is no time to wait for blood, the next thing is to stop the blood loss. This is done by means of a Sengstaken-Blakemore tube, a three lumened tube with oesophageal and gastric balloons. The tube should not be left in situ for more than three days, since sloughing of the mucosa will then occur.

A large quantity of blood has been lost into the gastrointestinal tract, and this is liable to give rise to hepatic coma due to reabsorption of nitrogenous products from the gut. As much of this as possible is removed via the gastric tube and by enema, and 1 gram of Neomycin Sulphate should be given in an attempt to reduce nitrogenous metabolism of the gut flora.

Once resuscitated, the patient will have to undergo full investigation in order to decide future management—surgery or not.

We have now to consider the cold case—who has probably had one or two haematemeses, who is, at the moment, in fairly good general condition but whose future is, to say the least, doubtful. Into this category we may put our resuscitated patient mentioned above. We have come to the fork in the road—medical or surgical treatment?—and we do not know which way to turn. If we ask the sympathetic onlooker which way to go he will smile and say he's not very sure, and if we consult our map, it will show us that both roads come to an end not very far hence.

Let us first consider the lessons learnt from 15 years of surgery in the portal system. In the first year after operation there is, in fact, a marked reduction in the incidence of subsequent haemorrhage as compared with medical series. Thereafter there is little to choose, but perhaps a slight advantage exists in surgical treatment. The surgical approach to patients with advanced liver disease has met with uniformly bad results. Perhaps a look at the criteria which must be satisfied before operation is undertaken will help us to decide what to do:—

(1) Age—patients over 60 years of age do not do well with surgery.

(2) Recent jaundice—contra-indication.

(3) Active liver disease—a contra-indication.
(4) Liver function tests—must show:
Serum albumin: not less than 3 gm. %.
Flocculation tests: if positive, must be only weakly so.
Bromsulphalein: must be excreted to 70% within 30 minutes.
(5) Ascites and Oedema—which fail to respond to medical treatment are strong contra-indications.

If our patient does not fit these rather stringent criteria, then medical treatment along the lines I have described should be instituted.

The basis of definite surgical treatment is the creation of a porto-systemic anastomosis, thus allowing decompression of the portal system Blakemore, the pioneer in this field, enunciated in 1945 three principles which still hold good:

(1) That shunt operations are designed to treat haemorrhage, and not ascites.
(2) That shunts must be large, the only useful veins for the purpose being the portal and splenic veins.
(3) That a shunt will function better the nearer it is to the obstruction.

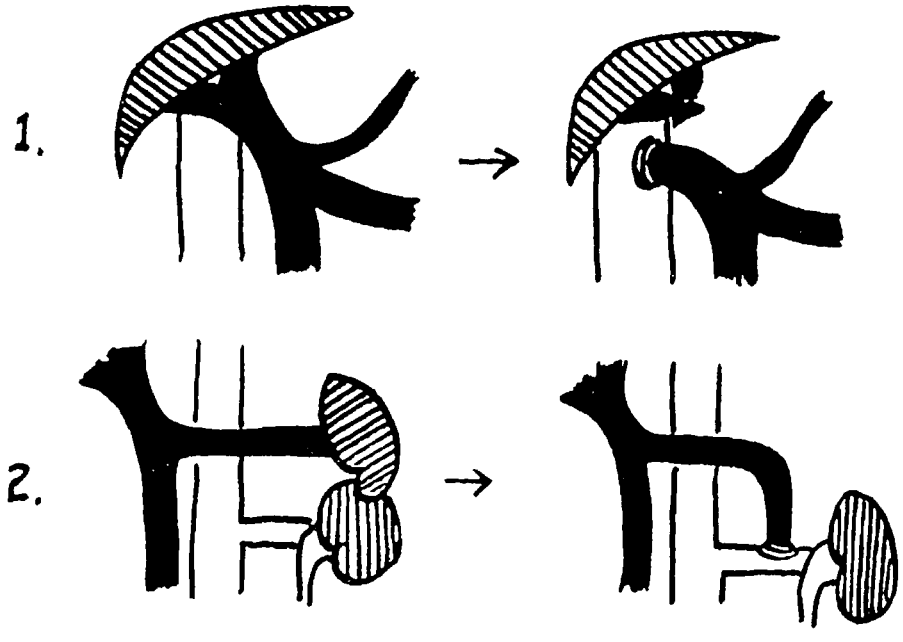

Fig. 3

The Portacaval Shunt

This is undoubtedly the most efficient anastomosis, but that brings us up against the big problem of shunts straight away—the more efficient the shunt the more likely is neuropathy to develop. However, in view of the less satisfying results obtained from other procedures, it seems reasonable to hold to Milnes Walker's dictum that, having decided a shunt is necessary, the most efficient anastomosis possible should be created. The mechanics

of the anastomosis are shown in figure 3. As a decompressing procedure this works admirably, but it seems to me to be basically wrong to effect a total diversion of the portal blood (as in end-to-side anastomosis) from an organ which has a precarious oxygen supply at the best of times. The alternative of side-to-side anastomosis does not solve the problem, since on the reduction of pressure the flow of blood upwards from the site of junction to the liver becomes either non-existent or even reversed, a state of affairs that is conducive to thrombosis. It is a remarkable fact that all attempts at manometry have been made with the patient in the horizontal position, and it would be interesting to measure what effect the erect posture has upon the pressure gradient across an anastomosis,

In cases where hypersplenism is present in sufficient degree to incapacitate the patient some workers advocate preliminary splenectomy, which is often followed by a reversion to normal of the blood dyscrasia. If this is done it is imperative to ensure that the portal vein is capable of being used later as a shunt, since the alternative splenorenal anastomosis can no longer be done. In these cases I believe the Edinburgh practise is to perform a splenorenal anastomosis in the first instance.

The Splenorenal Shunt

Originally Blakemore used to remove the left kidney, but obviously this is unnecessary (see figure 3). The indications for splenorenal anastomosis at the present time are:

1. The indications for portacaval shunting, with
2. an unsuitable portal vein, or
3. severe hypersplenism, and
4. a large splenic vein.

It is a more formidable procedure, and less efficient as a shunt, and for this reason it was at one time advocated as the operation of choice on the basis that it produced the required decompression with minimal neuropathic symptoms, whilst conserving the normal hepatic inflow. However, because of the difficulty of thrombosis, this is no longer the operation of choice without specific indications.

Operations on the Varices

There remain a number of operations directed at the varices themselves, and which are resorted to when no suitable vein for anastomosis exists, or when neuropathy is a feature of the patient's illness. There are many such operations, and I propose merely to list them with a few comments here and there:

1. Injection via the oesophagoscope. This merely destroys valuable porto-systemic connections, and raises the pressure in those that remain.
2. Gastric transection.
3. Oesophageal transection with splenectomy.
4. Ligature of the varices as an emergency procedure.
5. Limited Oesophago-gastrectomy of use when other operations have been or cannot be attempted.
6. Omentopexy—worthless.
7. Ligature of the hepatic and other (splenic) arteries has been advocated in Russia in preference to shunt operations for patients with cirrhosis, but after a few unsuccessful trials has commanded no great popularity in the West.

All these methods fail to prevent the recurrence of bleeding, later if

not sooner. Linton's idea that emergency ligature of the varices should give 3-4 weeks in which to improve the patient's condition prior to anastomosis seems sound, but even he himself did not suggest it as definative treatment.

Portal Neuropathy: Theories

As I mentioned earlier, the great difficulty of portal decompression is the likelihood of producing neuropathy by shunting the unfinished work of absorptive metabolism into the systemic circulation where the rough rocks of protein breakdown are crashed upon the gravel path of cerebral metabolism. Or so it seems, for even yet we are not certain of the real cause of portal neuropathy. A rather inconstant relation with arterial ammonia concentration has been demonstrated, and patients being treated with Neomycin may suffer a rise in the blood ammonia level without clinical deterioration. It seems certain, however, that nitrogenous materials are causally related, since the two main precipitating factors are hæmorrhage into the gastro-intestinal tract, and a heavy protein meal, but what these materials are, and what is their relationship to intestinal, liver and cerebral metabolism, nobody is yet prepared to say. Keto acids and amines have been accused, but not as yet proved guilty. There still remains the alternative that portal encephalopathy may be due to a deficiency, rather than an excess, of some factor: 5-hydroxy trpytophane has been suggested as a possibility in this sense.

Results of Surgical Treatment

The operative mortality has been variously reported from 9-15%, being lowest in splenorenal anastomosis because that operation is often done in cases of extrahepatic obstruction without gross liver damage. The main causes of fatality, according to Blakemore, are:

1. Hepatic failure and neuropathy.
2. Operative hæmorrhage.
3. Mesenteric thrombosis.
4. Recurrent hæmorrhage.

A good demonstration of the superiority of portacaval anastomosis over the splenorenal type is given in Eckman's figures, in which 14 out of 32 patients with splenorenal anastomosis had recurrence of bleeding, while no recurrence at all was observed in 20 patients with a portacaval shunt, who were followed up for between one and seven years.

The functional result is on the whole good, and it has been demonstrated here in Edinburgh that liver function is only temporarily depressed after anastomosis, and that no more than after gastrectomy. However, I would question anyone who asserted that *no* liver damage occurred in man: permanent damage certainly ensues in dogs subjected to a similar insult.

Prognosis

In his classical series on the dietary treatment of cirrhosis, Patek concluded that 18% of all cirrhotics died of gastro-intestinal hæmorrhage, and that about 50% of patients who bled were dead within a year of their first bleed. Other writers give different results, some more and others less favourable, but even the most optimistic figures demonstrate the hopelessness of this condition 20 years ago, and though we have come a long way on the road to success, this is no time to rest on our laurels. It appears that in this condition, as in so many others, we must turn to the biochemist and the serologist to unlock for us doors that are as yet closed.

[For References, see page 46

His fourth publication on renal disease—the Gulstonian lecture of 1833—presented a summary of the signs he had come to associate with albuminous urine—anasarca, uræmia, absence of urea from the urine, infections, cardiac hypertrophy, cerebral symptoms and pathology of the kidneys.

Bright died in 1838 at the height of a widespread and just reputation. His contribution to all branches of clinical research had been prolific—the heart, the liver, the spleen, the pancreas, the Gastro-intestinal tract, and the Central Nervous System—all had been faithfully observed in their pathological behaviour in life and appearance after death. He was a cheerful and attractive personality always careful to acknowledge the assistance of his juniors. In his heyday he had had no competition in research into kidney disease: the scope and detail of his own findings hardly encouraged it. But at the time of his death he had been retired from Guy's for 15 years, and would-be competitors, having sat back to take stock of the whole new concept, had in the meantime set the ball rolling again. So nephritis left Bright behind and emerged into the present century, where the successive efforts of Volhard and Fahr, Dorothy Russell and Ellis to classify his disease have detracted nothing from, and added many complications to, the original simple account of Richard Bright himself.

REFERENCES

1. Osman: Original Papers of Richard Bright on Bright's Disease.
2. Major: Bulletin of Medical History, 23, 453-460.
3. Guy's Hospital Reports, 1927.
4. Guy's Hospital Reports, 1958.
5. Hale-White: Great Doctors of the 19th century, 1935.
6. Nixon: Bright's Disease, 1954.

PORTAL HYPERTENSION (continued from page 20)

REFERENCES

1. Macpherson, A.I.S., Postgraduate Medical Journal, 1956, 32, 474.
2. Daniell and Pritchard, "J. Physiol." 1951, 114, 521.
3. Riddell *et al.* Clin. Sc., 1957, 16, 315.
4. Atkinson and Sherlock, "Lancet," 1954, 1, 1325.
5. Paton, Reynolds and Sherlock, "Lancet," 1953, 1, 918.
6. Cameron and De Saram, "J. Path. Bact.," 1939, 48, 41.
7. R. Milnes Walker, "Portal Hypertension," Edward Arnold, 1959.
8. Sherlock, "Diseases of the Liver," Livingstone, 1955.
9. Shobinger, Cooper and Rousselot, "Ann. Surg." 1959, 150, 188.
10. R. M. Walker, Proc. R. Soc. Med., 1943, 36, 445.
11. Atkinson, "Lancet," 1959, 2, 819.
12. Macpherson, Postgraduate Medical Journal, 1954, 30, 184.
13. R. M. Walker, "J. Fac. Radiol." 1957, 8, 178.
14. Hunt, Jacksonian Prize Essay, Royal College Surgeons, 1956.
15. Macbeth, "Brit. Med. J.," 1955, 2, 877.
16. Tanner, Ann. R. Coll. Surg., 1958, 22, 30.
17. Linton and Elliss, J. Amer. Med. Ass., 1956, 160, 1017.
18. Madden, Surg. Gynaec. Obstet., 1954, 99, 385.
19. Ryss, S. M., and Smagin, V. G., Terap, Arkh., 1958, 2, 37.
20. Dawson, McLaren, Sherlock, "Lancet," 1957, 2, 1263.
21. Macpherson, A.I.S., *et al*, "Lancet," 1954, 2, 353.
22. Large, *et al*, "Ann. Surg.," 1952, 135, 22.
23. Patek *et al*, J. Amer. Med. Ass., 1948, 138, 543.